CONGENITAL DEFORMITIES OF MECHANICAL ORIGIN*

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This and the following paper are both on subjects belonging to that strangely neglected group of human ills, the congenital deformities. To compare this group with others of similar importance, for instance infections or new growths, is to find that there is no generally accepted classification of its elements, and no textbook treating of them. What information is available has to be extracted from papers scattered through the literature and a few monographs; there is no general review of the subject.

The first lecture indeed treats of a group of deformities which is not generally admitted to exist at all, those due to mechanical faults in the mechanically extremely complicated process of gestation. It is strange that though it is universally admitted that mechanical faults can have their obvious consequences in the cavities of the skull or the chest, it is almost universally denied that similar processes and results may occur in the cavity of the uterus. One explanation of this may be that the experimental method, on which modern medicine relies almost entirely, is of little use here: it is too difficult to alter intra-uterine conditions without bringing the pregnancy to an end. In consequence the chief method available is one much used in such scientific pursuits as astronomy, that of working out in the imagination what would happen were certain hypotheses true, and then comparing these abstract conclusions with what actually occurs. The difference from the experimental method, in which actual changes are made in the course of events (for instance, Duraswami's researches on the effect of insulin on the developing egg), is fundamental, so that it is all the more strange that there is no term in common use to describe and distinguish this method of using the mind.

Abnormal Mechanics in Pregnancy

It is possible to imagine the process of gestation going wrong mechanically in three main ways:

Malposition. By this term I mean that the developing foetus, which must obviously be tightly folded up, is folded up in the wrong way. Here one comes against another interesting gap in present teaching; the lack of any generally accepted view on how the foetus is normally folded. Keith (1948), for instance, teaches that the feet lie *in utero* with their soles against each other, but this does not explain the usual excess of movement into calcaneus that a newborn baby's foot possesses over that of an adult. If we presume that the normal intra-uterine position is with the soles of the feet against the uterine wall and, of course, with the hips, knees, and elbows flexed, then this hypothesis is confirmed by the fact that the range of movement in the joints of a newborn infant corresponds to what would be expected from a prolonged sojourn in this position. That is, the elbows, knees, and hips will not extend to the adult range, and the feet go up into calcaneus much further, so that the little toes can easily be made to touch the outer side of the leg.

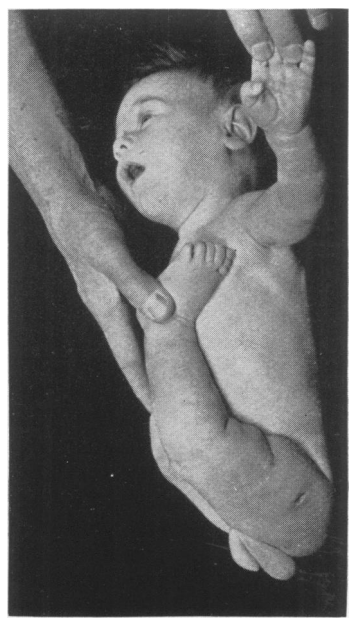

FIG. 1.—Example of malposition *in utero*. The pregnancy was normal in size and sensations, and the baby showed no evidence of excessive pressure. The only abnormality is that the knees are extended instead of flexed.

I do not know any orthopaedic work which comments on these facts, which is perhaps to be accounted for by most orthopaedic authorities at present being committed to a dogmatic denial

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that pathological conditions of the feet and joints can have mechanical causes. To admit that in the newborn there are normal variations from the adult state of obvious mechanical origin would be to allow the entry of the thin end of a heretical wedge-shaped idea.

Increased Mechanical Pressure. It might be assumed that increased pressure on a limb would have the same effects on the tissues before birth as it would have afterwards. If a plaster case is put too tightly on a limb the effects are two: atrophy of the muscles and stiffening of the joints. Probably the main cause of these effects is interference with the venous return, but the explanation of the stiffening of the joints is not obvious.

It might also be expected that if there were undue tightness of the uterus on the foetus, which should be the only cause of this pressure, there would be two effects felt on the maternal side. (1) The mother's abdomen would be smaller than in normal pregnancies. (2) There would be discomfort for the mother, particularly noticeable when she has had a normal pregnancy to compare with the abnormally tight one.

On checking this hypothesis with what actually occurs, it is found to be constantly confirmed: that is to say that children showing evidences of pressure in atrophied muscles and stiff joints emerge from small and uncomfortable pregnancies. The legs and feet are, of course, much more affected than the arms, which are sheltered by the large overhanging foetal head.

Increased Hydraulic Pressure. An increase in the tension of the amniotic fluid might be expected to interfere with the venous returns from the limbs just as a similar increase in the intracranial tension spoils the venous return from the retina and causes optic atrophy. The difference in the effects of this pressure from those of the mechanical type would be that owing to the laws of hydraulics it would be equally distributed over all four limbs. Its effects would also be expected to be more severe as the distance from the pump of the foetal heart becomes greater, that is to say more in the feet than in the hips, and more in the hands than in the shoulders.

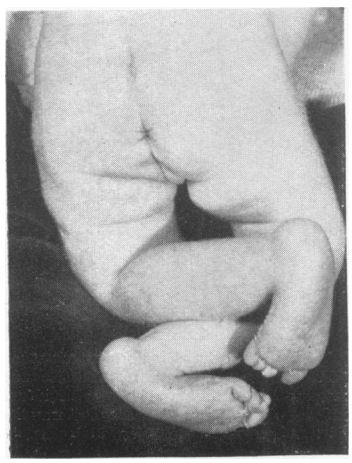

FIG. 2.—Example of excessive mechanical pressure. The pregnancy was a very small and uncomfortable one. The feet and legs have been in the normal position, but have been moulded to conform closely to the concavity of the uterus. The joints of the legs are stiff, and the muscles have hardly developed at all.

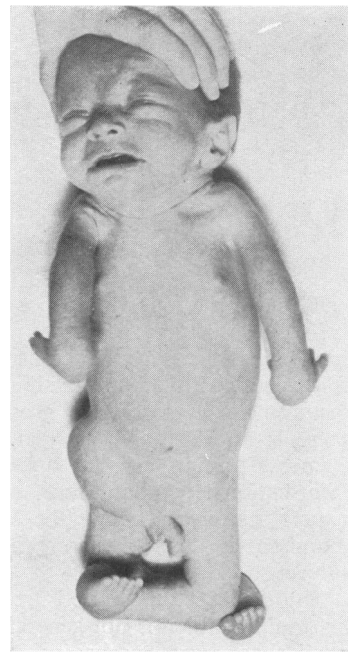

FIG. 3.—Example of increased hydraulic pressure. The pregnancy was a very large and uncomfortable one, with excessive amniotic fluid. The fixed stiff extension of the hips could only occur in a uterine cavity larger than normal. Note also the cylindrical limbs, showing none of the normal moulding of the tissues over the bony points, owing to lack of contact with the uterine wall. The joints are stiff, and the muscles undeveloped, equally in all four limbs owing to the laws of hydraulics, a contrast to the normal arms in Fig. 2.

Increased hydraulic pressure suggests that usually there would be an abnormally large amount of fluid; though, of course, it would be possible to have increased hydraulic pressure with a normal or abnormally small amount. Compare again the situation in the cranial cavity, where increased cerebral tension may exist either with the increased amount of fluid of hydrocephalus, or the decreased amount found in cerebral tumours.

Here again the maternal histories of cases of 'arthrogryposis' confirm the hypothesis. The mothers give a story of abnormal pain of the kind associated with abnormal hydraulic pressure in the skull, the bladder, or a joint. A very large proportion of these children emerge from hydramniotic pregnancies, and they often show the joints of the limbs stiffened in positions which would only be possible in an abnormally large cavity: for example, the hips are extended instead of flexed in the normal way.

It is characteristic of the attitude of present-day

medicine towards this subject of congenital deformities that the correlation of abnormal babies with abnormal pregnancies is almost entirely neglected. The reason is that the obstetrician is very little interested in the baby, and the paediatrician equally uninterested in the details of pregnancy, while the orthopaedic surgeon is hampered by the dogmatic attitude which I have already mentioned.

Classification of Deformities due to Mechanical Conditions

Deformities of the Feet. There are three regular deformities which occur continually in different degrees of the same moulding.

METATARSAL VARUS. This condition is due to mal-position with the soles of the feet together. There is also a form which is combined with metatarsal valgus of the other foot in a mutual deformity, which is assumed to the feet being diverted sideways in the normal position.

CLUB-FOOT, EQUINO-VARUS OR TOTAL VARUS. I object to the name equino-varus as in the more severe degrees the feet are in calcaneus, and I suggest the name 'total varus' to describe this condition in which the ankle and heel are moulded inwardly as well as the forefoot. The deformity can be most shortly described by saying that it is exactly what would result were one to press the outer side of the model of a foot in putty against a concavity corresponding to that of the uterine wall.

METATARSAL VALGUS. This is not due to malposition, but to increased mechanical pressure exaggerating the normal calcaneus position of the newborn foot so that it wraps itself round the outer side of the leg. Its association with congenital dislocation of the hip and weak muscles is mentioned later.

In addition to these regular types of deformity there is an infinite variety of irregular forms of moulding, corresponding to various abnormal positions of the limbs. These are associated in particular with spina bifida in those cases in which very early pressure has both interfered with the fusing of the edges of the spinal groove and forced the cerebellum and medulla down through the foramen magnum into the deformity known as the Arnold-Chiari malformation.

It is of interest to compare with the mechanical hypothesis the explanations usually given of feet mouldings of the type vaguely known as talipes.

That they are due to arrest at some stage of development: no one could possibly accept this hypothesis who is familiar with the infinite variety of mouldings of the feet, though at one stage there is a vague likeness between the inversion of the foetal feet and the mildest form of 'equino-varus' talipes (in the severer forms it has been noted that the feet described in this way are in calcaneus, not equinus).

That the deformities are due to the action of abnormal or abnormally attached muscles: apart from the fact that many of the mouldings could not possibly be produced in this way there is the awkward fact that some of the most severe of them occur in children whose muscles can never have been active, cases of spina bifida and arthrogryposis.

That club-foot or total varus is due to a congenital dislocation of the astragalo-scaphoid joint (Brockman, 1930): it is of course impossible to mould a foot into this shape without dislocating slightly this joint as well as others. But this explanation leaves unaccounted for the moulding of the heel and the atrophy of the calf muscles, quite apart from its offering no explanation of the immense variety of other malformations of the same type.

That 'the deformity is not a result of the position, but the position is a result of the deformity': This is a purely verbal objection, as can be seen by transposing it into the statement that 'the deformity of bow-legs is not a result of the position of bow-legs, but the position of bow-legs is a result of the deformity of bow-legs'.

The interesting experiments of Duraiswami (1952) have been accepted in some quarters as suggesting a pathology of talipes and congenital dislocation of the hip. Apart from the fact that such conditions often occur in perfectly healthy children, the deformities produced in chickens have only the vaguest resemblance to those found in man. The resemblance is much more a matter of words than of structure.

Deformities of the Joints. The only joint to be found dislocated in otherwise normal children is the hip, and it is the only joint that could be dislocated by mechanical means given the intra-uterine conditions. Also it is always dislocated in the only direction in which it could be forced, that is to say backwards and downwards by pressure on the knee; and the head and neck of the femur are compressed and anteverted in the way in which a soft structure like the foetal head of the femur would be moulded by being forced out of the socket while the joint is in the intra-uterine position.

The only other joint to be found dislocated, the knee, is only so malformed in association with the malposition of hyperextension, the connexion being very obvious.

Deformities of Other Parts. With the exception of the Arnold-Chiari deformity which has already been briefly mentioned, these are infinite in variety and irregular in type, very often being associated with embryological faults of formation such as missing digits and bones.

An important part of the argument is the combination of different deformities. To take a few of them:

In double talipes the deformity is never exactly equal, but always greater on one foot than the other. This would be expected as the outer foot in the cross-legged position responsible would get more moulding pressure than the inner one.

Congenital dislocation of the hip is constantly associated with valgus talipes. The foot deformity is assumed to be due to increased mechanical pressure, which would also tend by its action on the knee to push the head of the femur out backwards.

Congenital dislocation of the hip is never, in my experience, associated with a straightforward varus moulding of the foot. This is because the pressure on the outside of the foot swings the knee into abduction, which brings the head of the femur forward and keeps it in the acetabulum.

Metatarsal valgus talipes, assumed to be due to increased mechanical pressure, is always associated with weak muscles, which would also be expected from the same cause.

The existence of 'mutual deformities' in which the convexity of an irregular talipes of one foot fits into the concavity of an irregular talipes of the other. It seems to me that the occurrence of such cases would be alone sufficient to prove the moulding hypothesis.

Treatment

The essential idea of the classical treatment which I was originally taught was to treat deformities rather as if they had been cement or plaster, that is to say they were corrected as far as possible, tendons being freely cut to allow this, and they were then immobilized with the idea that they should set in the position obtained. This notion ignores the fact that the body is a vital moving structure, which only develops properly if its parts are being used in the way for which they are fitted. I found its results to be so disappointing that I sought for new principles, and found them in the idea that certain movements which tend to correct the deformity should be permitted, and others which increase it should be prevented. To do this it was necessary to work out a new system of splinting, the main idea of which was to control the system of levers, which is formed by bones, joints and muscles, by means of a counter-balancing system of levers in the form of splints.

The disadvantage of the old method was that the joints stiffened and the muscles atrophied under immobilization and the effects of cutting tendons round the ankle were much the same as those of cutting them round the wrist.

The advantages to be expected from the system of controlled movement would be:

(1) Rapidity of correction, due to the continual manipulations supplied by the automatic actions of the infant; (2) stimulation of the growth of imperfectly developed parts, a stimulation that use alone could give; (3) the acquiring of correct muscle balance by use in the corrected position. Immobilization, of course, simply weakens all the muscles equally, without changing their original faulty relationships of tone.

I have worked upon the idea that to reverse the moulding of the feet one should imitate the kind of pressure that has originally produced it. This pressure has certain characteristics:

(1) It is constant, so that manipulations at intervals do not reproduce it. (2) It is fluctuating, varying *in utero* with the motions of the child and of the uterus. This fluctuation can be imitated by a mechanism that allows movements, even though the movements are selected. One very important result of these fluctuations in pressure is that they tend to pump blood through the tissues, and so prevent that stasis of the circulation which is a main factor in pressure sores, caused either by immobilization in plaster or by prolonged lying still in bed. (3) It is soft and distributed over a wide area. This can be imitated by such material as sticking plaster or adhesive felt. (4) It is frictionless. In the uterus the amniotic fluid prevents friction. In splinting this can also be done by stopping the movement of the material which is applying pressure, that movement which is the essence of friction and consequent soreness, by making this material adhesive. It is quite an important point in technique which is often not understood, that if one uses adhesive felt to apply pressure it should be stuck to the foot, and not to the splint.

It is, of course, impossible to describe here the various forms of apparatus devised for the various deformities, but in my opinion the hopes I held of this method of treatment have been very fairly realized. Of course, some varieties of these deformities are utterly intractable; nothing can produce muscles which have failed to form, and I have never had any real success in diminishing the stiffness of congenitally stiff joints.

It is worth recording, not as a complaint but as

an observation of an instance of a universal human reaction, that I had the utmost difficulty originally in getting my heretical ideas of pathology and treatment published. This was because the editors of journals submitted my articles to the opinion of orthopaedic authorities, who, finding their teachings assailed, naturally said that the attack should not be allowed to go forth to corrupt the young.

At present my main trouble is due to another universal reaction, that of modifying a method or a mechanism in order to establish the superiority of the modifier over the original inventor. It is still apparently difficult for many surgeons to understand the system of levers of which my splints consist, as is shown by the curiously common cutting off of the extremely important outside lever in my 'hobble splint' for talipes.

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